A steroid free maintenance regimen is efficacious and safe for ANCA associated vasculitis and glomerulonephritis.

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Introduction

Corticosteroids (CS) are the mainstay of treatment in patients with ANCA associated vasculitis (AAV) but are associated with significant adverse effects. We report the successful induction treatment of patients with severe AAV using an extreme CS minimisation regimen.

Methods

Patients with active MPO- or PR3-ANCA vasculitis or ANCA negative, pauci-immune glomerulonephritis (GN) included. Induction treatment consisted intravenous methylprednisolone, 2 doses of rituximab, 3 months of low dose cyclophosphamide, and only 2 weeks of oral CS. We compared our patient outcomes with a matched cohort entered in EUVAS trials (CYCAZAREM, CYCLOPS, MEPEX, IMPROVE) with conventional steroid dosing.

Results

23 patients included with 6-month follow-up data, 21 patients at 12 months. 5 PR-3-ANCA, 16 MPO-ANCA and 2 ANCA negative. 22/23 patients with de novo renal disease, 1 with extra-renal relapse. 20/22 underwent renal biopsy, classified as: focal (8), crescentic (5), mixed (5), sclerosed (2). 4 subjects had pulmonary haemorrhage, 3 required dialysis.

Median prednisolone duration 15 days (IQR 14-27 days). Combining methylprednisolone and oral CS, median cumulative dose 1235mg (range 280-3940). 5 subjects had >30 days of CS: 3 due to protocol deviation, 2 due to on-going activity. All patients entered complete remission within 3 months.

Median baseline creatinine 209 μmol/L (range 89-999) by 12 months 109 μmol/L (range 64-216)($p<0.005$). Baseline median urine PCR 198 mg/mmol (range 11-611), by 12 months 29 mg/mmol (0-333) ($p<0.001$). Significant decreases in ANCA, baseline median titre 100 IU/ml (range 0-143) and at 12 months 3 IU/ml (0-134)($p<0.005$), as well as BVAS comparing baseline to all subsequent time-points (all $p<0.001$).

Three patients required dialysis (for 11, 36 and 53 days) before recovering independent function. No patients developed end-stage renal failure. 1 extra-renal relapse. No new cases of diabetes, no significant weight gain. Two patients had serious infections, 8 minor.

Compared to the matched EUVAS trials cohort (n=128), no difference in remission induction, change in GFR over six months, change in BVAS or CRP. Total steroid exposure significantly less 1235mg v 3847mg ($p<0.05$).

A national questionnaire demonstrated that usual practice in the UK is to continue steroids for more than six months with 24% never attempting any steroid withdrawal.

Conclusion

These data demonstrate that minimizing steroid exposure is safe in AAV. Results in this pilot study are comparable to trial data with traditional CS regimes.